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Search Results -

Term	Documents
(3 AND 1).USPT,JPAB,EPAB,DWPI,TDBD.	18

Database: US Patents Full-Text Database; JPO Abstracts Database; EPO Abstracts Database; Derwent World Patents Index; IBM Technical Disclosure Bulletins

Refine Search: L1 and L3 Clear

Search History

Today's Date: 8/17/2000

DB Name	Query	Hit Count	Set Name
USPT,JPAB,EPAB,DWPI,TDBD	L1 and L3	18	L8
USPT,JPAB,EPAB,DWPI,TDBD	L5 and L3	0	L7
USPT,JPAB,EPAB,DWPI,TDBD	L5 and L2	0	L6
USPT,JPAB,EPAB,DWPI,TDBD	L1 and L4	4	L5
USPT,JPAB,EPAB,DWPI,TDBD	(knockout mutation)	45	L4
USPT,JPAB,EPAB,DWPI,TDBD	(spinal muscular) adj (atrophy)	194	L3
USPT,JPAB,EPAB,DWPI,TDBD	(Smn gene)	9	L2
USPT,JPAB,EPAB,DWPI,TDBD	(transgenic mouse)	2613	L1

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Search Results - Record(s) 1 through 9 of 9 returned.

☑ 1. Document ID: US 6080577 A

L2: Entry 1 of 9 File: USPT Jun 27, 2000

US-PAT-NO: 6080577
DOCUMENT-IDENTIFIER: US 6080577 A
TITLE: Survival motor neuron (SMN) gene: a gene for spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

☐ 2. Document ID: US 5935783 A

L2: Entry 2 of 9 File: USPT Aug 10, 1999

US-PAT-NO: 5935783
DOCUMENT-IDENTIFIER: US 5935783 A
TITLE: Genes mapping in the digeorge and velocardiofacial syndrome minimal critical region

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

☐ 3. Document ID: US 5882868 A

L2: Entry 3 of 9 File: USPT Mar 16, 1999

US-PAT-NO: 5882868
DOCUMENT-IDENTIFIER: US 5882868 A
TITLE: Method of diagnosing spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

☐ 4. Document ID: JP 08228785 A

L2: Entry 4 of 9 File: JPAB Sep 10, 1996

PUB-NO: JP408228785A
DOCUMENT-IDENTIFIER: JP 08228785 A
TITLE: SURVIVAL MOTOR NEURON (SMN) GENE: SPIRAL MUSCULAR ATROPHY

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

---

☐ 5. Document ID: EP 711833 A2

L2: Entry 5 of 9 File: EPAB May 15, 1996

PUB-NO: EP000711833A2
DOCUMENT-IDENTIFIER: EP 711833 A2
TITLE: Survival motor neuron (SMN) gene: a gene for spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

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☐ 6. Document ID: EP 708178 A1

L2: Entry 6 of 9 File: EPAB Apr 24, 1996

PUB-NO: EP000708178A1
DOCUMENT-IDENTIFIER: EP 708178 A1
TITLE: Survival motor neuron (SMN) gene: a gene for spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

---

☑ 7. Document ID: AU 9964696 A, WO 200021553 A2, EP 999270 A1

L2: Entry 7 of 9 File: DWPI May 1, 2000

DERWENT-ACC-NO: 2000-317843
DERWENT-WEEK: 200036

TITLE: Use of interferons for increasing interferon regulatory factor or stat factor to regulate the survival motor neuron gene expression, and treating spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

---

☐ 8. Document ID: EP 711833 A2

L2: Entry 8 of 9 File: DWPI May 15, 1996

DERWENT-ACC-NO: 1996-232098
DERWENT-WEEK: 199624

TITLE: Human survival motor neuron gene T-BCD541, variant C-BCD541 and murine equiv. - useful to develop primers and probes for in vitro detection of motor neuron diseases e.g. spinal muscular atrophy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Image

☑ 9. Document ID: US 6080577 A, EP 708178 A1, AU 9534369 A, CA 2160937 A, JP 08228785 A, EP 711833 A3, AU 702252 B

L2: Entry 9 of 9 File: DWPI Jun 27, 2000

DERWENT-ACC-NO: 1996-202055
DERWENT-WEEK: 200036

TITLE: New isolated survival motor neuron gene - used to develop prods. for the diagnosis and treatment of motor neuron diseases

Full | Title | Citation | Front | Review | Classification | Date | Reference | Claims | KWIC | Draw Desc | Clip Img | Image

Generate Collection

Term	Documents
SMN.DWPI,TDBD,EPAB,JPAB,USPT.	226
SMNS	0
GENE.DWPI,TDBD,EPAB,JPAB,USPT.	120974
GENES.DWPI,TDBD,EPAB,JPAB,USPT.	36178
(SMN ADJ GENE).USPT,JPAB,EPAB,DWPI,TDBD.	9

Display 10 Documents, starting with Document: 9

Display Format: TI Change Format

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### Status: Path 1 of [Dialog Information Services via Modem]

### Status: Initializing TCP/IP using (UseTelnetProto 1 ServiceID pto-dialog)
Trying 3106900061...Open

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### Status: Signing onto Dialog
 ********
ENTER PASSWORD:
 ******** HHHHHHHH SSSSSSSS? ********
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### Status: Connected


Dialog level 00.07.20D

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Logon file001 17aug00 10:04:59
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***WIPO/PCT Patents Fulltext (File 349)

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***Datamonitor Market Research (File 761)
***Dissertation Abstracts Online (File 35)
***GPO Monthly Catalog (File 66)
***Bridge World Markets News (File 609,809)
***Fort Worth Star-Telegram (File 427)
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RELOADED
***Canadian Business Directory (File 533)
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***D&B European Dun's Market Identifiers (File 521)
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***CANCERLIT (File 159)

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File   1:ERIC  1966-2000/Jul 26
       (c) format only 2000 The Dialog Corporation

      Set  Items  Description
      ---  -----  -----------
?b 155,5,73
       17aug00 10:05:15 User259876 Session D101.1
            $0.42    0.120 DialUnits File1
     $0.42  Estimated cost File1
     $0.01  TYMNET
     $0.43  Estimated cost this search
     $0.43  Estimated total session cost   0.120 DialUnits
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SYSTEM:OS  - DIALOG OneSearch
  File 155:MEDLINE(R)  1966-2000/Oct W1
         (c) format only 2000 Dialog Corporation
  File   5:Biosis Previews(R)  1969-2000/Aug W3
         (c) 2000 BIOSIS
  File  73:EMBASE  1974-2000/Jul W4
         (c) 2000 Elsevier Science B.V.
*File  73: Update codes are currently undergoing readjustment.
For details type Help News73.

      Set  Items  Description
      ---  -----  -----------
?s (SMN (w) gene?)
Processing
             991  SMN
         5347215  GENE?
      S1     409  (SMN (W) GENE?)
?s (spinal (w) muscular (w) atrophy)
          348407  SPINAL
          267218  MUSCULAR
          109072  ATROPHY
      S2    4026  (SPINAL (W) MUSCULAR (W) ATROPHY)
?s (transgenic (w) (mouse or mice))
           76050  TRANSGENIC
         1181644  MOUSE
         1071999  MICE
      S3   40099  (TRANSGENIC (W) (MOUSE OR MICE))
?s s1 and s2 and s3
             409  S1
            4026  S2
           40099  S3
      S4       1  S1 AND S2 AND S3
?t s4/3,k/all

 4/3,K/1     (Item 1 from file: 5)
DIALOG(R)File   5:Biosis Previews(R)
(c) 2000 BIOSIS. All rts. reserv.

12137256   BIOSIS NO.: 199900432105
Creation and characterization of *transgenic* *mice* expressing Cre
 recombinase in skeletal muscle and neurons.
AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a);
  Dierich A(a); le Meur M(a); Melki J(a)
AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg**France
1999
JOURNAL: European Journal of Human Genetics  7 (SUPPL. 1):p113-114 July,
1999
CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human
Genetics  Geneva, Switzerland  May 29-June 1, 1999
ISSN: 1018-4813
RECORD TYPE: Citation
LANGUAGE: English

Creation and characterization of *transgenic* *mice* expressing Cre
 recombinase in skeletal muscle and neurons.
DESCRIPTORS:
  DISEASES: *spinal* *muscular* *atrophy*--
  CHEMICALS & BIOCHEMICALS:   ...murine *SMN* *gene* (Muridae...
?ds

Set     Items   Description
S1        409   (SMN (W) GENE?)
S2       4026   (SPINAL (W) MUSCULAR (W) ATROPHY)
S3      40099   (TRANSGENIC (W) (MOUSE OR MICE))
```

```
S4          1   S1 AND S2 AND S3
?s s1 and s3
            409  S1
          40099  S3
      S5      1  S1 AND S3
?t s5/3,k/all

 5/3,K/1     (Item 1 from file: 5)
DIALOG(R)File   5:Biosis Previews(R)
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12137256   BIOSIS NO.: 199900432105
Creation and characterization of *transgenic* *mice* expressing Cre
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AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a);
  Dierich A(a); le Meur M(a); Melki J(a)
AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg**France
1999
JOURNAL: European Journal of Human Genetics  7 (SUPPL. 1):p113-114 July,
1999
CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human
Genetics  Geneva, Switzerland  May 29-June 1, 1999
ISSN: 1018-4813
RECORD TYPE: Citation
LANGUAGE: English

Creation and characterization of *transgenic* *mice* expressing Cre
 recombinase in skeletal muscle and neurons.
DESCRIPTORS:
  CHEMICALS & BIOCHEMICALS:   ...murine *SMN* *gene* (Muridae...
?ds

Set     Items   Description
S1        409   (SMN (W) GENE?)
S2       4026   (SPINAL (W) MUSCULAR (W) ATROPHY)
S3      40099   (TRANSGENIC (W) (MOUSE OR MICE))
S4          1   S1 AND S2 AND S3
S5          1   S1 AND S3
?s s2 and s3
            4026  S2
           40099  S3
      S6      21  S2 AND S3
?rd
...completed examining records
      S7      16  RD (unique items)
?s s7 and (human (w) SMN (w) gene)
Processing
              16  S7
        15705149  HUMAN
             991  SMN
         1496029  GENE
              40  HUMAN(W)SMN(W)GENE
      S8       0  S7 AND (HUMAN (W) SMN (W) GENE)
?s s7 and (knockout (w) mutation)
              16  S7
           19660  KNOCKOUT
          402502  MUTATION
             305  KNOCKOUT(W)MUTATION
      S9       0  S7 AND (KNOCKOUT (W) MUTATION)
?s s1 and (transgenic (w) animal?)
Processing
             409  S1
           76050  TRANSGENIC
        14521177  ANIMAL?
            6613  TRANSGENIC(W)ANIMAL?
     S10       0  S1 AND (TRANSGENIC (W) ANIMAL?)
```

?s s1 and (transgenic (w) rat?)
Processing
409 S1
76050 TRANSGENIC
6228380 RAT?
1427 TRANSGENIC(W)RAT?
S11 0 S1 AND (TRANSGENIC (W) RAT?)
?t s7/3,k/all

7/3,K/1 (Item 1 from file: 155)
DIALOG(R)File 155:MEDLINE(R)

10367554 20122241
The human centromeric survival motor neuron gene (SMN2) rescues embryonic lethality in Smn(-/-) mice and results in a mouse with *spinal* *muscular* *atrophy*.
Monani UR; Sendtner M; Coovert DD; Parsons DW; Andreassi C; Le TT; Jablonka S; Schrank B; Rossol W; Prior TW; Morris GE; Burghes AH
Departments of Neurology, College of Medicine, Ohio State University, Columbus, OH 43210, USA.
Human molecular genetics (ENGLAND) Feb 12 2000, 9 (3) p333-9, ISSN 0964-6906 Journal Code: BRC
Contract/Grant No.: NS38650, NS, NINDS
Languages: ENGLISH
Document type: JOURNAL ARTICLE

The human centromeric survival motor neuron gene (SMN2) rescues embryonic lethality in Smn(-/-) mice and results in a mouse with *spinal* *muscular* *atrophy*.
Proximal *spinal* *muscular* *atrophy* (SMA) is a common motor neuron disease in humans and in its most severe form causes death by the age of 2 years. It is...
... SMN2). Mice possess only one survival motor neuron gene (Smn) whose loss is embryonic lethal. Therefore, to obtain a mouse model of SMA we created *transgenic* *mice* that express human SMN2 and mated these onto the null Smn (-/-)background. We show that Smn (-/-); SMN2 mice carrying one or two copies of the...
Chemical Name: Nerve Tissue Proteins; (SMN protein (*spinal* *muscular* *atrophy*)

7/3,K/2 (Item 2 from file: 155)
DIALOG(R)File 155:MEDLINE(R)

10264509 20082811
A mouse model for *spinal* *muscular* *atrophy*.
Hsieh-Li HM; Chang JG; Jong YJ; Wu MH; Wang NM; Tsai CH; Li H
Institute of Molecular Biology, Academia Sinica, Taipei, Taiwan.
Nature genetics (UNITED STATES) Jan 2000, 24 (1) p66-70, ISSN 1061-4036 Journal Code: BRO
Languages: ENGLISH
Document type: JOURNAL ARTICLE

A mouse model for *spinal* *muscular* *atrophy*.
... motor neuron gene is present in humans in a telomeric copy, SMN1, and several centromeric copies, SMN2. Homozygous mutation of SMN1 is associated with proximal *spinal* *muscular* *atrophy* (SMA), a severe motor neuron disease characterized by early childhood onset of progressive muscle weakness. To understand the functional role of SMN1 in SMA, we produced mouse lines deficient for mouse Smn and *transgenic* *mouse* lines that expressed human SMN2. Smn-/- mice died during the peri-implantation stage. In contrast, *transgenic* *mice* harbouring SMN2 in the Smn-/- background showed pathological changes in the spinal cord and skeletal muscles similar to those of SMA patients. The severity of...

Chemical Name: DNA Primers; (Nerve Tissue Proteins; (SMN protein (*spinal* *muscular* *atrophy*)

7/3,K/3 (Item 3 from file: 155)
DIALOG(R)File 155:MEDLINE(R)

08567047 97173353
Animal models of ALS.
Pioro EP; Mitsumoto H
Cleveland Clinic Foundation, OH 44195, USA.
Clinical neuroscience (UNITED STATES) 96 1995, 3 (6) p375-85, ISSN 1065-6766 Journal Code: B9U
Languages: ENGLISH
Document type: JOURNAL ARTICLE; REVIEW; REVIEW, TUTORIAL

... models have been most extensively studied, including three mouse models: motor neuron degeneration (Mnd), progressive motor neuronopathy (pmn), wobbler, and one canine model: hereditary canine *spinal* *muscular* *atrophy* (HCSMA). The wobbler mouse has been the most extensively studied of these models with analyses of clinical, pathological (perikaryon, axon, muscle), and biochemical features. Experimentally...

... mouse models in which genes relevant to the human disease or motor neuron biology have been manipulated. The most clinically relevant of these is a *transgenic* *mouse* overexpressing the mutated SOD1 gene of FALS patients, which has already provided significant insights into mechanisms of motor neuron degeneration in this disease. Because no...

7/3,K/4 (Item 1 from file: 5)
DIALOG(R)File 5:Biosis Previews(R)

12214817 BIOSIS NO.: 199900509666
Creation and characterization of *transgenic* *mouse* lines expressing Cre recombinase in skeletal muscle or neurons.
AUTHOR: Tiziano F(a); Miniou P(a); Frugier T(a); Dierich A(a); Le Meur M(a) ; Melki J(a)
AUTHOR ADDRESS: (a)IGBMC. INSERM/CNRS/ULP, Illkirch, Strasbourg**France 1999
JOURNAL: American Journal of Human Genetics 65 (4):pA493 Oct., 1999
CONFERENCE/MEETING: 49th Annual Meeting of the American Society of Human Genetics San Francisco, California, USA October 19-23, 1999
SPONSOR: The American Society of Human Genetics
ISSN: 0002-9297
RECORD TYPE: Citation
LANGUAGE: English

Creation and characterization of *transgenic* *mouse* lines expressing Cre recombinase in skeletal muscle or neurons.
DESCRIPTORS:
DISEASES: *spinal* *muscular* *atrophy*--

7/3,K/5 (Item 2 from file: 5)
DIALOG(R)File 5:Biosis Previews(R)

12137256 BIOSIS NO.: 199900432105
Creation and characterization of *transgenic* *mice* expressing Cre recombinase in skeletal muscle and neurons.
AUTHOR: Tiziano Francesco(a); Miniou P(a); Frugier T(a); Roblot N(a); Dierich A(a); le Meur M(a); Melki J(a)
AUTHOR ADDRESS: (a)IGBMC, Illkirch, Strasbourg**France

1999
JOURNAL: European Journal of Human Genetics 7 (SUPPL. 1):p113-114 July, 1999
CONFERENCE/MEETING: 31st Annual Meeting of the European Society of Human Genetics Geneva, Switzerland May 29-June 1, 1999
ISSN: 1018-4813
RECORD TYPE: Citation
LANGUAGE: English

Creation and characterization of *transgenic* *mice* expressing Cre recombinase in skeletal muscle and neurons.
DESCRIPTORS:
DISEASES: *spinal* *muscular* *atrophy*--

7/3,K/6 (Item 1 from file: 73)
DIALOG(R)File 73:EMBASE

07515254 EMBASE No: 1998422152
The genetic and molecular mechanisms of motor neuron disease
Wong P.C.; Rothstein J.D.; Price D.L.
P.C. Wong, Department of Pathology, The Johns Hopkins University, School of Medicine, 720 Rutland Avenue, Baltimore, MD 21205-2196 United States
AUTHOR EMAIL: pcwong@welchlink.welch.jhu.edu
Current Opinion in Neurobiology (CURR. OPIN. NEUROBIOL.) (United Kingdom) 1998, 8/6 (791-799)
CODEN: COPUE ISSN: 0959-4388
DOCUMENT TYPE: Journal; Review
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 84

MEDICAL DESCRIPTORS:
gene isolation; *transgenic* *mouse*; gene targeting; clinical feature; spastic paraplegia--congenital disorder--cn; *spinal* *muscular* *atrophy* --congenital disorder--cn; human; review; priority journal

7/3,K/7 (Item 2 from file: 73)
DIALOG(R)File 73:EMBASE

07477051 EMBASE No: 1998395122
Pathogenesis of neurodegenerative diseases associated with expanded glutamine repeats: New answers, new questions
Ross C.A.; Margolis R.L.; Becher M.W.; Wood J.D.; Engelender S.; Cooper J.K.; Sharp A.H.
C.A. Ross, Johns Hopkins University, School of Medicine, Department of Psychiatry, 720 Rutland Avenue, Baltimore, MD 21205-2196 United States
AUTHOR EMAIL: caross@jhu.edu
Progress in Brain Research (PROG. BRAIN RES.) (Netherlands) 1998, 117/- (397-419)
CODEN: PBRRA ISSN: 0079-6123
DOCUMENT TYPE: Journal; Conference Paper
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 155

...polar zipper', into beta pleated sheets. Recent findings have now established the presence of such aggregates in selected regions of brain from affected individuals, in *transgenic* *mice* expressing expanded repeats, and in isolated cells transfected with expanded repeats. The aggregates are most prominently manifest as neuronal intranuclear inclusion bodies. As the investigation...
MEDICAL DESCRIPTORS:
*huntington chorea--etiology--et; *hereditary *spinal* *muscular* *atrophy* --etiology--et; *trinucleotide repeat

neurologic disease--etiology--et; degenerative disease--etiology--et; *spinal* *muscular* *atrophy*--etiology--et; *transgenic* *mouse*; gene deletion; gene mutation; genetic predisposition; neurotoxicity--etiology --et; human; nonhuman; mouse; animal model; controlled study; human tissue; animal tissue; conference paper; priority journal

7/3,K/8 (Item 3 from file: 73)
DIALOG(R)File 73:EMBASE

07273542 EMBASE No: 1998185616
Androgen receptor YAC *transgenic* *mice* carrying CAG 45 alleles show trinucleotide repeat instability
La Spada A.R.; Peterson K.R.; Meadows S.A.; McClain M.E.; Jeng G.; Chmelar R.S.; Haugen H.A.; Chen K.; Singer M.J.; Moore D.; Trask B.J.; Fischbeck K.H.; Clegg C.H.; McKnight G.S.
A.R. La Spada, Box 357110, University Washington Medical Center, 1959 NE Pacific Street, Seattle, WA 98195-7110 United States
AUTHOR EMAIL: laspada@mail.labmed.washington.edu
Human Molecular Genetics (HUM. MOL. GENET.) (United Kingdom) 1998, 7/6 (959-967)
CODEN: HMGEE ISSN: 0964-6906
DOCUMENT TYPE: Journal; Article
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 42

Androgen receptor YAC *transgenic* *mice* carrying CAG 45 alleles show trinucleotide repeat instability

...alleles (37-66 CAGs) change in length when transmitted from parents to offspring, with a significantly greater tendency to shift size when inherited paternally. As *transgenic* *mice* carrying human AR cDNAs with 45 and 66 CAG repeats do not display repeat instability, we attempted to model trinucleotide repeat instability by generating *transgenic* *mice* with yeast artificial chromosomes (YACs) carrying AR CAG repeat expansions in their genomic context. Studies of independent lines of AR YAC *transgenic* *mice* with CAG 45 alleles reveal intergenerational instability at an overall rate of ~ 10%. We also find that the 45 CAG repeat tracts are significantly more unstable with maternal transmission and as the transmitting mother ages. Of all the CAG/CTG repeat *transgenic* *mice* produced to date the AR YAC CAG 45 mice are unstable with the smallest trinucleotide repeat mutations, suggesting that the length threshold for repeat instability...
MEDICAL DESCRIPTORS:
*trinucleotide repeat; *hereditary *spinal* *muscular* *atrophy* --congenital disorder--cn
yeast artificial chromosome; allele; X chromosome linkage; exon; *transgenic* *mouse*; dna flanking region; dna sequence; sequence tagged site; gene mapping; gene locus; polymerase chain reaction; parental age; immunoblotting; fluorescence in situ hybridization; nonhuman; mouse; animal ...

7/3,K/9 (Item 4 from file: 73)
DIALOG(R)File 73:EMBASE

07247164 EMBASE No: 1998136081
Evidence of mdx mouse skeletal muscle fragility in vivo by eccentric running exercise
Vilquin J.-T.; Brussee V.; Asselin I.; Kinoshita I.; Gingras M.; Tremblay J.P.
Dr. J.P. Tremblay, Centre de Recherche en Neurobiologie, Univ. Laval and Hopital Enfant-Jesus, 1401, 18e Rue, Laval, Que. G1J IZ4 Canada
Muscle and Nerve (MUSCLE NERVE) (United States) 1998, 21/5 (567-576)

CODEN: MUNED ISSN: 0148-639X
DOCUMENT TYPE: Journal; Article
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 51

...of 18 months, and the role of dystrophin in fiber integrity is not fully understood. The fragility of the skeletal muscle fibers was investigated in *transgenic* *mice* expressing beta-galactosidase under the control of a muscle specific promoter. Adult mdx/beta-galactosidase (dystrophinnegative) and normal/beta-galactosidase (dystrophin-positive) mice were submitted...
MEDICAL DESCRIPTORS:
running; exercise; hereditary *spinal* *muscular* *atrophy*--diagnosis--di; hereditary *spinal* *muscular* *atrophy*--etiology--et; enzyme analysis; protein expression; phenotype; nonhuman; male; female; mouse; animal model; controlled study; animal tissue; animal cell; article; priority journal

7/3,K/10 (Item 5 from file: 73)
DIALOG(R)File 73:EMBASE

07084073 EMBASE No: 1997365936
***Transgenic* *mouse* models of neurodegenerative disease caused by CAG/polyglutamine expansions**
Bates G.P.; Davies S.W.
Dr. G.P. Bates, Medical and Molecular Genetics, UMDS, Guy's Hospital, London SE1 9RT United Kingdom
AUTHOR EMAIL: g.bates@umds.ac.uk
Molecular Medicine Today (MOL. MED. TODAY) (United Kingdom) 1997, 3/11 (508-518)
CODEN: MMTOF ISSN: 1357-4310
PUBLISHER ITEM IDENTIFIER: S1357431097011428
DOCUMENT TYPE: Journal; Review
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 47

***Transgenic* *mouse* models of neurodegenerative disease caused by CAG/polyglutamine expansions**
MEDICAL DESCRIPTORS:
animal model; cell death; gene expression; gene mutation; genetic stability ; huntington chorea--etiology--et; machado joseph disease--etiology--et; molecular genetics; mouse; nonhuman; phenotype; review; *spinal* *muscular* *atrophy*--etiology--et; spinocerebellar degeneration--etiology--et; *transgenic* *mouse*

7/3,K/11 (Item 6 from file: 73)
DIALOG(R)File 73:EMBASE

06965185 EMBASE No: 1997249782
The CAG/polyglutamine tract diseases: Gene products and molecular pathogenesis
Koshy B.T.; Zoghbi H.Y.
Dr. B.T. Koshy, Howard Hughes Medical Institute, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030 United States
Brain Pathology (BRAIN PATHOL.) (United States) 1997, 7/3 (927-942)
CODEN: BRPAE ISSN: 1015-6305
DOCUMENT TYPE: Journal; Conference Paper
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 157

...polyglutamine confers a gain of function onto the involved protein. To understand the mechanisms underlying the pathogenesis of these diseases, investigators have turned to generating *transgenic* *mice* which

recapitulate some of the features of the human disease and hence are excellent model systems to study the progression of the disease in vivo.
MEDICAL DESCRIPTORS:
*fragile X syndrome--etiology--et; **spinal* *muscular* *atrophy*--etiology --et; *spinocerebellar degeneration--etiology--et

7/3,K/12 (Item 7 from file: 73)
DIALOG(R)File 73:EMBASE

06725751 EMBASE No: 1997007213
Glutamine repeats and inherited neurodegenerative diseases: Molecular aspects
Perutz M.F.
M.F. Perutz, MRC Laboratory of Molecular Biology, Hills Road, Cambridge CB2 2QH United Kingdom
Current Opinion in Structural Biology (CURR. OPIN. STRUCT. BIOL.) (United Kingdom) 1996, 6/6 (848-858)
CODEN: COSBE ISSN: 0959-440X
DOCUMENT TYPE: Journal; Review
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH
NUMBER OF REFERENCES: 62

...41 repeats. Protein constructs with more than 41 repeats are toxic to E. coli and to CHO cells in culture, and they elicit ataxia in *transgenic* *mice*. These observations argue in favour of a distinct change of structure associated with elongation beyond 37-41 glutamine repeats. The review describes experiments designed to...
MEDICAL DESCRIPTORS:
ataxia; cho cell; cytotoxicity; dentatorubropallidoluysian atrophy --etiology--et; dentatorubropallidoluysian atrophy--congenital disorder--cn ; dna sequence; escherichia coli; hereditary *spinal* *muscular* *atrophy* --etiology--et; hereditary *spinal* *muscular* *atrophy*--congenital disorder--cn; human; huntington chorea--congenital disorder--cn; huntington chorea--etiology--et; hydrogen bond; machado joseph disease--congenital disorder--cn; machado joseph disease--etiology--et; mouse; nonhuman; oligomerization; priority journal; protein conformation; protein folding; review; spinocerebellar degeneration--congenital disorder--cn; spinocerebellar degeneration--etiology--et; structure activity relation; *transgenic* *mouse*

7/3,K/13 (Item 8 from file: 73)
DIALOG(R)File 73:EMBASE

06716854 EMBASE No: 1996381838
Animal models for motor neuron diseases: Research directions
Ludolph A.C.
Department of Neurology, University of Ulm, Steinhovelstrasse 9,89075 Ulm Germany
Neurology (NEUROLOGY) (United States) 1996, 47/6 SUPPL. 4 (S228-S232)
CODEN: NEURA ISSN: 0028-3878
DOCUMENT TYPE: Journal; Review
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

...neuron degenerative, wobbler, wasted, and autosomal-recessive progressive motor neuropathy mouse models), sporadically occurring (e.g., equine motor neuron disease), and experimentally induced (e.g., *transgenic* *mice* carrying a mutation in the gene encoding human Cu/Zn superoxide dismutase, toxic models) disorders. The models currently have three major drawbacks. First, there is...
MEDICAL DESCRIPTORS:
animal model; anterior horn cell disease--etiology--et; anterior horn cell disease--diagnosis--di; gene mutation; hereditary *spinal* *muscular*

atrophy--diagnosis--di; hereditary *spinal* *muscular* *atrophy* --etiology--et; lipofuscinosis--etiology--et; lipofuscinosis--diagnosis--di ; nonhuman; priority journal; pyramidal tract; review; *transgenic* *mouse*

7/3,K/14 (Item 9 from file: 73)
DIALOG(R)File 73:EMBASE

06529216 EMBASE No: 1996194816
Transgenic and gene-targeting approaches to model disorders of motor neurons
Wong P.C.; Borchelt D.R.; Lee M.K.; Pardo C.A.; Sisodia S.S.; Cleveland D.W.; Koliatsos V.E.; Price D.K.
Department of Pathology, Johns Hopkins Univ. School Medicine,Baltimore, MD United States
Seminars in the Neurosciences (SEMIN. NEUROSCI.) (United Kingdom) 1996 , 8/3 (163-169)
CODEN: SNEUE ISSN: 1044-5765
DOCUMENT TYPE: Journal; Article
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

...Over the past several years, significant progress has been made in understanding the genetics of some of these disorders, including familial amyotrophic lateral sclerosis (FALS), *spinal* *muscular* *atrophy* (SMA), and spinal bulbar muscular atrophy (SBMA). For example, some of the autosomal dominant cases of FALS are linked to mutations in the superoxide dismutase 1 (SOD1) gene. Several groups have introduced these SOD1 mutations into *transgenic* *mice*, and these animals develop features of the human disease. Other investigators have used transgenic strategies to overexpress wild-type (wt) or mutant neurofilament (NF) genes...
MEDICAL DESCRIPTORS:
...sclerosis--etiology--et; animal experiment; animal model; article; cell death; cell inclusion; cytoskeleton; gene mutation; hyperreflexia; mouse; muscle atrophy; muscle weakness; neurofilament; nonhuman; phenotype; spasticity; *spinal* *muscular* *atrophy*--etiology--et; *transgenic* *mouse*

7/3,K/15 (Item 10 from file: 73)
DIALOG(R)File 73:EMBASE

06217067 EMBASE No: 1995245815
Genetic approaches to pathogenesis of neurodegenerative diseases
Orr H.T.; Clark H.B.
Laboratory Medicine/Pathology Dept., Institute of Human Genetics, University of Minnesota,Minneapolis, MN United States
Laboratory Investigation (LAB. INVEST.) (United States) 1995, 73/2 (161-171)
CODEN: LAINA ISSN: 0023-6837
DOCUMENT TYPE: Journal; Review
LANGUAGE: ENGLISH

MEDICAL DESCRIPTORS:
...etiology--et; machado joseph disease--etiology--et; major clinical study ; male; nerve cell necrosis; neurologic disease--etiology--et; neuropathology; nonhuman; onset age; priority journal; review; *spinal* *muscular* *atrophy*--etiology--et; *transgenic* *mouse*

7/3,K/16 (Item 11 from file: 73)
DIALOG(R)File 73:EMBASE

06019153 EMBASE No: 1995049283

Stability of an expanded trinucleotide repeat in the androgen receptor gene in *transgenic* *mice*

Bingham P.M.; Scott M.O.; Wang S.; McPhaul M.J.; Wilson E.M.; Garbern J.Y.; Merry D.E.; Fischbeck K.H.
Division of Neurology, Children's Hospital of Philadelphia,Philadelphia, PA 19104 United States
Nature Genetics (NAT. GENET.) (United States) 1995, 9/2 (191-196)
CODEN: NGENE ISSN: 1061-4036
DOCUMENT TYPE: Journal; Article
LANGUAGE: ENGLISH SUMMARY LANGUAGE: ENGLISH

Stability of an expanded trinucleotide repeat in the androgen receptor gene in *transgenic* *mice*

...stability of a trinucleotide repeat and to develop an animal model of one of these disorders, spinal and bulbar muscular atrophy (SBMA), we have generated *transgenic* *mice* carrying either the normal or expanded repeat human androgen receptor (AR) gene. Unlike the disease allele in humans, the AR cDNA containing the expanded repeat in *transgenic* *mice* showed no change in repeat length with transmission. Expression of the SBMA AR was found in *transgenic* *mice*, but at a lower level than normal endogenous expression. The lack of a physiological pattern of expression may explain why no phenotypic effects of the...
MEDICAL DESCRIPTORS:
**spinal* *muscular* *atrophy*; *transgene
animal model; animal tissue; article; controlled study; densitometry; gene expression; mouse; nonhuman; phenotype; polyacrylamide gel electrophoresis; polymerase chain reaction; priority journal; southern blotting; *transgenic* *mouse*
?ds

Set	Items	Description
S1	409	(SMN (W) GENE?)
S2	4026	(SPINAL (W) MUSCULAR (W) ATROPHY)
S3	40099	(TRANSGENIC (W) (MOUSE OR MICE))
S4	1	S1 AND S2 AND S3
S5	1	S1 AND S3
S6	21	S2 AND S3
S7	16	RD (unique items)
S8	0	S7 AND (HUMAN (W) SMN (W) GENE)
S9	0	S7 AND (KNOCKOUT (W) MUTATION)
S10	0	S1 AND (TRANSGENIC (W) ANIMAL?)
S11	0	S1 AND (TRANSGENIC (W) RAT?)

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